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L3

24 L2 AND ("USH2A" OR "USH 2A" OR "USH2 A" OR "USH-2A")

L3 ANSWER 1 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2004:307918 BIOSIS  
DN PREV200400305710  
TI Genetic analysis of 2299delG and C759F mutations (**USH2A**) in  
patients with visual and/or auditory impairments.  
AU Aller, Elena; Najera, Carmen [Reprint Author]; Millan, Jose M.; Oltra,  
Juan S.; Perez-Garrigues, Herminio; Vilela, Concepcion; Navea, Amparo;  
Beneyto, Magdalena  
CS Fac Ciencias BiolDept Genet, Univ Valencia, Dr Moliner 50, E-46100,  
Valencia, Spain  
Carmen.Najera@uv.es  
SO European Journal of Human Genetics, (May 2004) Vol. 12, No. 5, pp.  
407-410. print.  
ISSN: 1018-4813.  
DT Article  
LA English  
ED Entered STN: 7 Jul 2004  
Last Updated on STN: 7 Jul 2004

L3 ANSWER 2 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2003:387003 BIOSIS  
DN PREV200300387003  
TI The molecular genetics of Usher syndrome.  
AU Ahmed, Z. M.; Riazuddin, S.; Wilcox, E. R. [Reprint Author]  
CS Laboratory of Molecular Genetics, Section on Human Genetics, NIDCD, NIH, 5  
Research Court, 2A-19, Rockville, MD, 20850-3227, USA  
wilcoxe@nidcd.nih.gov  
SO Clinical Genetics, (June 2003) Vol. 63, No. 6, pp. 431-444. print.  
ISSN: 0009-9163 (ISSN print).  
DT Article  
General Review; (Literature Review)  
LA English  
ED Entered STN: 20 Aug 2003  
Last Updated on STN: 20 Aug 2003

L3 ANSWER 3 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2003:28147 BIOSIS  
DN PREV200300028147  
TI **Usherin** expression is highly conserved in mouse and human  
tissues.  
AU Pearsall, Nicole [Reprint Author]; Bhattacharya, Gautam; Wisecarver, Jim;  
Adams, Joe; Cosgrove, Dominic; Kimberling, William  
CS Boys Town National Research Hospital, 555 No. 30th St., Omaha, NE, USA  
kimber@boystown.org  
SO Hearing Research, (December 2002) Vol. 174, No. 1-2, pp. 55-63. print.  
ISSN: 0378-5955 (ISSN print).  
DT Article  
LA English  
ED Entered STN: 1 Jan 2003  
Last Updated on STN: 1 Jan 2003

L3 ANSWER 4 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2002:23328 BIOSIS  
DN PREV200200023328  
TI Distribution of **usherin** in humans and its effects on  
reproduction in people with usher syndrome type II.  
AU Pearsall, N. A. [Reprint author]; Bhattacharya, G. [Reprint author];  
Cosgrove, D. [Reprint author]; Wisecarver, J. L.; Kimberling, W. J.  
[Reprint author]  
CS Genetics Department, Boys Town National Research Hospital, Omaha, NE, USA

SO American Journal of Human Genetics, (October, 2001) Vol. 69, No. 4 Supplement, pp. 651. print.  
Meeting Info.: 51st Annual Meeting of the American Society of Human Genetics. San Diego, California, USA. October 12-16, 2001.  
CODEN: AJHGAG. ISSN: 0002-9297.

DT Conference; (Meeting)  
Conference; Abstract; (Meeting Abstract)  
Conference; (Meeting Poster)

LA English

ED Entered STN: 26 Dec 2001  
Last Updated on STN: 25 Feb 2002

L3 ANSWER 5 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2001:348191 BIOSIS  
DN PREV200100348191  
TI **Usherin** mutations associated with phenotypic variation in Usher syndrome type IIa and retinitis pigmentosa patients.  
AU Orten, D. J. [Reprint author]; Zeigler, T. [Reprint author]; Weston, M. D. [Reprint author]; Carney, C. A. [Reprint author]; Kimberling, W. J. [Reprint author]  
CS Boys Town National Research Hospital, Omaha, NE, USA  
SO IOVS, (March 15, 2001) Vol. 42, No. 4, pp. S644. print.  
Meeting Info.: Annual Meeting of the Association for Research in Vision and Ophthalmology. Fort Lauderdale, Florida, USA. April 29-May 04, 2001.  
Association for Research in Vision and Ophthalmology.  
DT Conference; (Meeting)  
Conference; Abstract; (Meeting Abstract)

LA English

ED Entered STN: 25 Jul 2001  
Last Updated on STN: 19 Feb 2002

L3 ANSWER 6 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2001:306545 BIOSIS  
DN PREV200100306545  
TI Spectrum of mutations in **USH2A** in British patients with Usher syndrome type II.  
AU Leroy, Bart P. [Reprint author]; Aragon-Martin, Jose A.; Weston, Michael D.; Bessant, David A. R.; Willis, Catherine; Webster, Andrew R.; Bird, Alan C.; Kimberling, William J.; Payne, Annette M.; Bhattacharya, Shomi S.  
CS Department of Molecular Genetics, Institute of Ophthalmology, 11-43 Bath Street, London, EC1V 9EL, UK  
bart.leroy@rug.ac.be  
SO Experimental Eye Research, (May, 2001) Vol. 72, No. 5, pp. 503-509. print.  
CODEN: EXERA6. ISSN: 0014-4835.  
DT Article  
LA English  
ED Entered STN: 27 Jun 2001  
Last Updated on STN: 19 Feb 2002

L3 ANSWER 7 OF 24 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2000:252758 BIOSIS  
DN PREV200000252758  
TI Genomic structure and identification of novel mutations in **Usherin**, the gene responsible for Usher syndrome type IIa.  
AU Weston, M. D.; Eudy, J. D.; Fujita, S.; Yao, S.-F.; Usami, S.; Cremers, C.; Greenburg, J.; Ramesar, R.; Martini, A.; Moller, C.; Smith, R. J.; Sumegi, J.; Kimberling, William J. [Reprint author]  
CS Boys Town National Research Hospital, 555 North 30th Street, Omaha, NE, 68131, USA  
SO American Journal of Human Genetics, (April, 2000) Vol. 66, No. 4, pp. 1199-1210. print.  
CODEN: AJHGAG. ISSN: 0002-9297.  
DT Article

LA English  
ED Entered STN: 21 Jun 2000  
Last Updated on STN: 5 Jan 2002

L3 ANSWER 8 OF 24 CANCERLIT on STN  
AN 2002168523 CANCERLIT  
DN 22106108 PubMed ID: 12112664  
TI Mutations in myosin VIIA (MYO7A) and usherin (USH2A)  
in Spanish patients with Usher syndrome types I and II, respectively.  
AU Najera Carmen; Beneyto Magdalena; Blanca Jose; Aller Elena; Fontcuberta  
Ana; Millan Jose Maria; Ayuso Carmen  
CS Departamento de Genetica, Facultad de Ciencias Biologicas, Universidad de  
Valencia, Valencia, Spain.. Carmen.Najera@uv.es  
SO HUMAN MUTATION, (2002 Jul) 20 (1) 76-7.  
Journal code: 9215429. ISSN: 1098-1004.  
CY United States  
DT Journal; Article; (JOURNAL ARTICLE)  
LA English  
FS MEDLINE; Priority Journals  
OS MEDLINE 2002366655  
EM 200208  
ED Entered STN: 20021018  
Last Updated on STN: 20021018

L3 ANSWER 9 OF 24 CAPLUS COPYRIGHT 2005 ACS on STN  
AN 2004:699598 CAPLUS  
DN 141:329966  
TI USH2A mutation analysis in 70 Dutch families with Usher syndrome  
type II  
AU Pennings, Ronald J. E.; te Brinke, Heleen; Weston, Michael D.; Claassen,  
Annemarie; Orten, Dana J.; Weekamp, Henriette; van Aarem, Annelies;  
Huygen, Patrick L. M.; Deutman, August F.; Hoefsloot, Lies H.; Cremers,  
Frans P. M.; Cremers, Cor W. R. J.; Kimberling, William J.; Kremer, Hannie  
CS Department of Otorhinolaryngology, UMC Nijmegen, Nijmegen, Neth.  
SO Human Mutation (2004), 24(2), 730/1-730/8  
CODEN: HUMUE3; ISSN: 1059-7794  
PB Wiley-Liss, Inc.  
DT Journal  
LA English

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L3 ANSWER 10 OF 24 CAPLUS COPYRIGHT 2005 ACS on STN  
AN 2004:694718 CAPLUS  
DN 141:275526  
TI Comprehensive screening of the USH2A gene in Usher syndrome type  
II and non-syndromic recessive retinitis pigmentosa  
AU Seyedahmadi, Babak Jian; Rivolta, Carlo; Keene, Julia A.; Berson, Eliot  
L.; Dryja, Thaddeus P.  
CS Harvard Medical School, Ocular Molecular Genetics Institute, Massachusetts  
Eye and Ear Infirmary, Boston, MA, 02114, USA  
SO Experimental Eye Research (2004), 79(2), 167-173  
CODEN: EXERA6; ISSN: 0014-4835  
PB Elsevier  
DT Journal  
LA English

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ALL CITATIONS AVAILABLE IN THE RE FORMAT

L3 ANSWER 11 OF 24 CAPLUS COPYRIGHT 2005 ACS on STN  
AN 2004:537880 CAPLUS  
DN 141:293912  
TI Immunohistochemistry and Reverse Transcriptase-Polymerase Chain Reaction

AU as Methods for Diagnostic Determination of Usher Syndrome Type IIa  
Cohn, Edward; Bhattacharya, Gautam; Pearsall, Nicole; Shendrik, Igor;  
Kimberling, William; Cosgrove, Dominic  
CS Usher Syndrome Center, Creighton University School of Medicine, Omaha, NE,  
USA  
SO Laryngoscope (2004), 114(7), 1310-1314  
CODEN: LARYA8; ISSN: 0023-852X  
PB Lippincott Williams & Wilkins  
DT Journal  
LA English  
RE.CNT 14 THERE ARE 14 CITED REFERENCES AVAILABLE FOR THIS RECORD  
ALL CITATIONS AVAILABLE IN THE RE FORMAT

L3 ANSWER 12 OF 24 CAPLUS COPYRIGHT 2005 ACS on STN  
AN 2002:603796 CAPLUS  
DN 137:308198  
TI Genetics of Usher-syndrome  
AU Bolz, Hanno; Gal, Andreas  
CS Institut fur Humangenetik des Universitatsklinikums, Hamburg-Eppendorf,  
Germany  
SO Medizinische Genetik (2002), 14(1), 10-14  
CODEN: MGENEZ; ISSN: 0936-5931  
PB Verlag Medizinische Genetik  
DT Journal; General Review  
LA German  
RE.CNT 9 THERE ARE 9 CITED REFERENCES AVAILABLE FOR THIS RECORD  
ALL CITATIONS AVAILABLE IN THE RE FORMAT

L3 ANSWER 13 OF 24 CAPLUS COPYRIGHT 2005 ACS on STN  
AN 2002:576854 CAPLUS  
DN 137:350607  
TI Mutations in myosin VIIA (MYO7a) and usherin (USH2a)  
in Spanish patients with Usher syndrome types I and II, respectively  
AU Najera, Carmen; Beneyto, Magdalena; Blanca, Jose; Aller, Elena;  
Fontcuberta, Ana; Millan, Jose Maria; Ayuso, Carmen  
CS Departamento de Genetica. Facultad de Ciencias Biologicas. Universidad de  
Valencia, Valencia, 46100, Spain  
SO Human Mutation (2002), 20(1), 513/1-513/7  
CODEN: HUMUE3; ISSN: 1059-7794  
PB Wiley-Liss, Inc.  
DT Journal  
LA English  
RE.CNT 39 THERE ARE 39 CITED REFERENCES AVAILABLE FOR THIS RECORD  
ALL CITATIONS AVAILABLE IN THE RE FORMAT

L3 ANSWER 14 OF 24 CAPLUS COPYRIGHT 2005 ACS on STN  
AN 2002:556025 CAPLUS  
DN 137:124204  
TI Usherin protein, gene, antibodies and immunoconjugates for  
diagnosis and therapy of Usher syndrome type IIa  
IN Cosgrove, Dominic E.  
PA Boys Town National Research Hospital, USA  
SO U.S. Pat. Appl. Publ., 40 pp.  
CODEN: USXXCO  
DT Patent  
LA English  
FAN.CNT 1

|      | PATENT NO.      | KIND | DATE     | APPLICATION NO. | DATE     |
|------|-----------------|------|----------|-----------------|----------|
| PI   | US 2002098516   | A1   | 20020725 | US 2001-970318  | 20011003 |
| PRAI | US 2000-237834P | P    | 20001003 |                 |          |

L3 ANSWER 15 OF 24 DGENE COPYRIGHT 2005 The Thomson Corp on STN

AN ABG32845 Protein DGENE  
TI Determining Usher syndrome type IIa in individual and detecting human  
usherin protein, by incubating sample with antibody  
immunoreactive with usher protein to produce immunoconjugate, and  
detecting immunoconjugate -  
IN Cosgrove D E  
PA (BOYS-N) BOYSTOWN NAT RES HOSPITAL.  
PI US 2002098516 A1 20020725 40  
AI US 2001-970318 20011003  
PRAI US 2000-237834P 20001003  
DT Patent  
LA English  
OS 2002-690477 [74]  
CR N-PSDB: ABS52998  
DESC Human **Usherin** protein **USH2a**.

L3 ANSWER 16 OF 24 DGENE COPYRIGHT 2005 The Thomson Corp on STN  
AN ABG32844 Protein DGENE  
TI Determining Usher syndrome type IIa in individual and detecting human  
usherin protein, by incubating sample with antibody  
immunoreactive with usher protein to produce immunoconjugate, and  
detecting immunoconjugate -  
IN Cosgrove D E  
PA (BOYS-N) BOYSTOWN NAT RES HOSPITAL.  
PI US 2002098516 A1 20020725 40  
AI US 2001-970318 20011003  
PRAI US 2000-237834P 20001003  
DT Patent  
LA English  
OS 2002-690477 [74]  
DESC Human **Usherin** protein **USH2a**, immunogenic peptide #2.

L3 ANSWER 17 OF 24 DGENE COPYRIGHT 2005 The Thomson Corp on STN  
AN ABG32843 Peptide DGENE  
TI Determining Usher syndrome type IIa in individual and detecting human  
usherin protein, by incubating sample with antibody  
immunoreactive with usher protein to produce immunoconjugate, and  
detecting immunoconjugate -  
IN Cosgrove D E  
PA (BOYS-N) BOYSTOWN NAT RES HOSPITAL.  
PI US 2002098516 A1 20020725 40  
AI US 2001-970318 20011003  
PRAI US 2000-237834P 20001003  
DT Patent  
LA English  
OS 2002-690477 [74]  
DESC Human **Usherin** protein **USH2a**, immunogenic peptide #1.

L3 ANSWER 18 OF 24 DGENE COPYRIGHT 2005 The Thomson Corp on STN  
AN ABS52998 cDNA DGENE  
TI Determining Usher syndrome type IIa in individual and detecting human  
usherin protein, by incubating sample with antibody  
immunoreactive with usher protein to produce immunoconjugate, and  
detecting immunoconjugate -  
IN Cosgrove D E  
PA (BOYS-N) BOYSTOWN NAT RES HOSPITAL.  
PI US 2002098516 A1 20020725 40  
AI US 2001-970318 20011003  
PRAI US 2000-237834P 20001003  
DT Patent  
LA English  
OS 2002-690477 [74]  
CR P-PSDB: ABG32845

DESC Human cDNA encoding **Usherin** protein **USH2a**.

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AN 2004153358 EMBASE  
TI Mutational spectrum in Usher syndrome type II.  
AU Ouyang X.M.; Yam D.; Hejtmancik J.F.; Jacobson S.G.; Li A.R.; Du L.L.;  
Angeli S.; Kaiser M.; Balkany T.; Liu X.Z.  
CS Dr. X.Z. Liu, Department of Otolaryngology, University of Miami, 1666 NW  
12th Avenue, Miami, FL 33136, United States. xliu@med.miami.edu  
SO Clinical Genetics, (2004) Vol. 65, No. 4, pp. 288-293.  
Refs: 32  
ISSN: 0009-9163 CODEN: CLGNAY  
CY United Kingdom  
DT Journal; Article  
FS 012 Ophthalmology  
022 Human Genetics  
LA English  
SL English  
ED Entered STN: 20040422  
Last Updated on STN: 20040422

L3 ANSWER 20 OF 24 FEDRIP COPYRIGHT 2005 NTIS on STN  
AN 2005:177436 FEDRIP  
NR CRISP 5R01DC004844-03  
TI **USHERIN: STRUCTURAL AND FUNCTIONAL ANALYSIS**  
SF Principal Investigator: COSGROVE, DOMINIC E; COSGROVE@BOYSTOWN.ORG, FATHER  
FLANAGAN'S BOYS' HOME, 555 NORTH 30TH STREET, OMAHA, NE 68131  
CSP FATHER FLANAGAN'S BOYS' HOME, BOYS TOWN, NEBRASKA  
CSS Supported By: NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION  
DISORDERS  
DB 2009 (/01/02)  
FYR 2004  
DE 2008 (/31/07)  
FU Noncompeting Continuation (Type 5)  
FS National Institutes of Health

L3 ANSWER 21 OF 24 GENBANK® COPYRIGHT 2005 on STN

LOCUS (LOC): AY481573 GenBank (R)  
GenBank ACC. NO. (GBN): AY481573  
GenBank VERSION (VER): AY481573.1 GI:44887472  
CAS REGISTRY NO. (RN): 664940-65-2  
SEQUENCE LENGTH (SQL): 18883  
MOLECULE TYPE (CI): mRNA; linear  
DIVISION CODE (CI): Primates  
DATE (DATE): 30 Mar 2004  
DEFINITION (DEF): Homo sapiens Usher syndrome 2A isoform B (**USH2A**)  
mRNA, complete cds.  
SOURCE: Homo sapiens (human)  
ORGANISM (ORGN): Homo sapiens  
Eukaryota; Metazoa; Chordata; Craniata; Vertebrata;  
Euteleostomi; Mammalia; Eutheria; Primates; Catarrhini;  
Hominidae; Homo  
REFERENCE: 1 (bases 1 to 18883)  
AUTHOR (AU): Van Wijk,E.; Pennings,R.J.; Te Brinke,H.; Claassen,A.;  
Yntema,H.G.; Hoefsloot,L.H.; Cremers,F.P.;  
Cremers,C.W.; Kremer,H.  
TITLE (TI): Identification of 51 Novel Exons of the Usher Syndrome  
Type 2A (**USH2A**) Gene That Encode Multiple  
Conserved Functional Domains and That Are Mutated in  
Patients with Usher Syndrome Type II  
JOURNAL (SO): Am. J. Hum. Genet., 74 (4), 738-744 (2004)

OTHER SOURCE (OS): CA 140:386911  
 REFERENCE: 2 (bases 1 to 18883)  
 AUTHOR (AU): van Wijk, E.; te Brinke, H.; Kremer, H.  
 TITLE (TI): Direct Submission  
 JOURNAL (SO): Submitted (19-NOV-2003) Otorhinolaryngology, UMC  
 Nijmegen, Geert Grootplein 10, Nijmegen 6525 GA,  
 Netherlands

FEATURES (FEAT):

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 Talmadge,C.B.; Pretto,D.; Weston,M.D.; Lehman,J.E.;  
 Zhou,M.; Seemayer,T.A.; Ahmad,I.; Kimberling,W.J.;  
 Sumegi,J.  
 TITLE (TI): Identification of the Mouse and Rat Orthologs of the  
 Gene Mutated in Usher Syndrome Type IIA and the  
 Cellular Source of **USH2A** mRNA in Retina, a  
 Target Tissue of the Disease  
 JOURNAL (SO): Genomics, 80 (2), 195-203 (2002)  
 OTHER SOURCE (OS): CA 137:347242  
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 AUTHOR (AU): Sumegi,J.; Huang,D.; Davis,J.R.  
 TITLE (TI): Direct Submission  
 JOURNAL (SO): Submitted (31-JAN-2002) Center for Human Molecular  
 Genetics, University of Nebraska Medical Center, 985454  
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GA The Genuine Article (R) Number: 911CZ  
TI Genetic and biochemical analyses of the **Ush2A** protein (  
usherin)  
AU Liu X (Reprint); Bulgakov O V; Pawlyk B; Adamian M; Li T  
SO INVESTIGATIVE OPHTHALMOLOGY & VISUAL SCIENCE, (2005) Vol. 46, Supp. [S].  
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TI Mutations in myosin VIIA (MYO7a) and usherin (USH2a)  
in Spanish patients with Usher syndrome types I and II, respectively  
AU Najera, Carmen; Beneyto, Magdalena; Blanca, Jose; Aller, Elena;  
Fontcuberta, Ana; Millan, Jose Maria; Ayuso, Carmen  
CS Departamento de Genetica. Facultad de Ciencias Biologicas. Universidad de  
Valencia, Valencia, 46100, Spain.  
SO Human Mutation, (2002) Vol. 20, No. 1, pp. 513/1-513/7.  
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L2 ANSWER 1 OF 11 BIOTECHDS COPYRIGHT 2005 THE THOMSON CORP. on STN  
DUPLICATE 1  
AN 2004-23454 BIOTECHDS  
TI Selectively transducing retinal pigment epithelium (RPE) cells using a  
vector particle exhibiting an AAV-4 capsid protein, useful for  
preventing, treating or alleviating an eye disease in a mammal;  
adeno-associated virus vector-mediated gene transfer and  
expression in host cell for eye disease gene therapy  
AU ROLLING F; WEBER M  
PA UNIV NANTES  
PI WO 2004084951 7 Oct 2004  
AI WO 2004-EP4020 26 Mar 2004  
PRAI US 2003-400531 28 Mar 2003; US 2003-400531 28 Mar 2003  
DT Patent  
LA English  
OS WPI: 2004-710276 [69]

L2 ANSWER 2 OF 11 IFIPAT COPYRIGHT 2005 IFI on STN DUPLICATE 2  
AN 10701606 IFIPAT;IFIUDB;IFICDB  
TI METHOD AND VECTORS FOR SELECTIVELY TRANSDUCING RETINAL PIGMENT EPITHELIUM

CELLS  
IN Rolling Fabienne (FR); Weber Michel (FR)  
PA Unassigned Or Assigned To Individual (68000)  
PI US 2004208847 A1 20041021  
AI US 2003-400531 20030328  
FI US 2004208847 20041021  
DT Utility; Patent Application - First Publication  
FS CHEMICAL  
APPLICATION  
CLMN 22  
GI 5 Figure(s).  
FIG. 1. Rat model: Rats were injected with rAAV-2/4. CMV.gfp and analyzed 30 days post injection (p.i.). Fluorescent retinal imaging (A). Sclera/choroid/RPE (B) and neuroretina (C) flatmounts. Sections from sclera/choroid/RPE (D) and neuroretina (E) examined under an inverted fluorescence microscope. RPE: retinal pigmented epithelium; ONL: outer nuclear layer; INL: inner nuclear layer; GCL: ganglion cell layer.  
FIG. 2. Nonhuman primate model: Live fluorescent retinal imaging at different time intervals (14, 21, 35, and 60 days p.i.) in Mac1 and Mac2. Both individuals received rAAV-2/4. CMV.gfp. (\*) retinal detachment created by the subretinal injection.  
FIG. 3. Nonhuman primate model: Two months p.i., neuroretina (A, B and D) and choroid/RPE (C) flatmounts were performed and examined under inverted fluorescence microscope. M, macula; ONH, optical nerve head, RV, retinal vessel.  
FIG. 4. Nonhuman primate model: Sections from neuroretina (A, B) and choroid/RPE (C, D) flatmounts and were either analyzed by normal light microscope (A, C) or inverted fluorescence microscope (B, D). See legend FIG. 1 for RPE, ONL, INL and GCL.  
FIG. 5. Vector shedding after subretinal delivery of rAAV-2/4. CMV.gfp in nonhuman primate (Mac1). PCR assay for sensitivity (A). Serum (s), lacrymal (I) and nasal (n) samples are represented (B). DNA marker (M), positive control on 25 pg of vector plasmid (+), negative control on water (-). Samples were collected 15 min, 2 hr and from day 1 to 28 p.i.

L2 ANSWER 3 OF 11 SCISEARCH COPYRIGHT (c) 2005 The Thomson Corporation on STN  
AN 2004:662568 SCISEARCH  
GA The Genuine Article (R) Number: 837QT  
TI Immunohistochemistry and reverse transcriptase-polymerase chain reaction as methods for diagnostic determination of Usher syndrome type IIa  
AU Cohn E; Bhattacharya G; Pearsall N; Shendrik I; Kimberling W; Cosgrove D (Reprint)  
CS Boys Town Natl Res Hosp, Usher Syndrome Ctr, 555 N 30th St, Omaha, NE 68131 USA (Reprint); Boys Town Natl Res Hosp, Usher Syndrome Ctr, Omaha, NE 68131 USA; Creighton Univ, Sch Med, Dept Pathol, Omaha, NE USA Cosgrove@boystown.org  
CYA USA  
SO LARYNGOSCOPE, (JUL 2004) Vol. 114, No. 7, pp. 1310-1314.  
ISSN: 0023-852X.  
PB LIPPINCOTT WILLIAMS & WILKINS, 530 WALNUT ST, PHILADELPHIA, PA 19106-3621 USA.  
DT Article; Journal  
LA English  
REC Reference Count: 14  
ED Entered STN: 13 Aug 2004  
Last Updated on STN: 13 Aug 2004  
\*ABSTRACT IS AVAILABLE IN THE ALL AND IALL FORMATS\*

L2 ANSWER 4 OF 11 IFIPAT COPYRIGHT 2005 IFI on STN DUPLICATE 3  
AN 10154875 IFIPAT;IFIUDB;IFICDB  
TI IMMUNODIAGNOSTIC DETERMINATION OF USHER SYNDROME TYPE IIa; DIAGNOSING PREFERENTIAL GENETIC DISORDER IN HUMANS; OBTAIN SAMPLE, INCUBATE WITH

ANTIBODY, DETECT BOUND ANTIBODY, COMPARE TO CONTROL, EVALUATE PATTERN FOR GENETIC DISORDER.

IN Cosgrove Dominic E  
PA Boys Town National Research Hospital (63491)  
PI US 2002098516 A1 20020725  
AI US 2001-970318 20011003  
PRAI US 2000-237834P 20001003 (Provisional)  
FI US 2002098516 20020725  
DT Utility; Patent Application - First Publication  
FS CHEMICAL  
APPLICATION  
OS CA 137:89726  
CLMN 41  
GI 9 Figure(s).

FIG. 1 illustrates the major structural elements of the **usherin** protein based on amino acid sequence. The amino acid positions where domains start and end are indicated. The location of polypeptides used to derive antibodies 1 (SEQ ID NO:1) and 2 (SEQ ID NO:2) used in these studies are shown. Constructs used to generate fusion peptides comprised the indicated portions of the LN, LE, and fibronectin type III domains (LN-FP, LE-FP, and FN-FP, respectively).

FIG. 2 is a Western blot of immunoprecipitated protein from extracts of retina and cochlea. For both gels: lane 1 is retinal extract; lane 2 is retinal extract immunoprecipitated with pre-immune serum; lane 3 is cochlear extract; lane 4 is cochlear extract precipitated with pre-immune serum. For the gel on the left, lanes 1 and 3 were immunoprecipitated with antibody 2 and blot probed with antibody 1. For the gel in the right, lane 1 and 3 were immunoprecipitated with antibody 1 and the blot was probed with antibody 2.

FIG. 3 is commercially available PolyA+ RNA dot blot from various mouse tissues. The blot was hybridized to a cDNA fragment corresponding to the LN domain of the protein. The template on the right indicates the tissues from which the corresponding RNA spot on the left was prepared.

FIG. 4 is an immunoperoxidase detection of tissues where **usherin** is **expressed**. A survey for **usherin expression** was conducted on mouse tissues. This figure summarizes where **usherin** was **expressed**. Serial sections were stained with hematoxylin and eosin (H&E) to illustrate tissue architecture, or with antiusherin (left panels), or anti-collagen alpha 1 (IV), which specifically localizes to the basement membranes. Arrows indicate **usherin** in the capillary basement membranes of the epididymus (D) and the spleen (J). Epidid=epididymus; Submax=submaxillary gland; Sm int=small intestine.

FIG. 5 is an immunoperoxidase detection of tissues where **usherin** is not **expressed**. Serial section were stained with hematoxylin and eosin (H&E) to illustrate tissue architecture, or with antiusherin (left panels), or anti-collagen alpha 1 (IV), which specifically localizes to the basement membranes. Sk musc=skeletal muscle; Sm musc=smooth muscle. Magnification bars are 50 mu m.

FIG. 6 is an **expression** of **usherin** in the inner ear and the eye of the mouse, and in the human retina. Mid-modiolar cross sections of the adult (8 wks) cochlea (A, B, C), or post-natal day 0 cochlea (G, H, I), or cross sections of adult retina (D, E, F) were immunostained with anti-**usherin** antibodies (A, D, G) or anti-type IV collagen antibodies (C, F, E). Eosin and hematoxylin stained serial sections are illustrated to provide a cellular frame of reference (B, E, H). Arrows in A and C denote the strial capillary basement membranes, and arrows in D, E, and F denote immunostaining in the basement membranes in Bruch's layer of the retina. Panel J shows **expression** of the **usherin** protein in the Bruch's layer and the choroid capillaries in human retina. Human retina was immunostained using the anti-**usherin** (raised against the mouse protein) antibody. Arrow heads indicate linear immunostaining in the

basement membranes on either limiting side of the Bruch's layer (BL). RPE=retinal pigment epithelial side; CL=choroid layer. Magnification bars are 50  $\mu$  m.

FIG. 7 is an immunogold localization of **usherin** to the basement membranes in striae capillaries, and the basement membrane in Bruch's layer of the retina. Arrows indicate immunogold particle deposition in the striae capillary basement membranes (A) and the basement membranes of the Bruch's layer (B) establishing **usherin** as a basement membrane protein. Note the proximity of the type I collagen fibrils with the basement membrane in B. CL=capillary lumen; MC=marginal cell; IPM=interphotoreceptor cell matrix; BL=Bruch's layer. Magnification bars are 50  $\mu$  m.

FIG. 8 is a Western blot illustrating the direct interaction of **usherin** with type IV collagen and the indirect interaction of **usherin** with type I collagen. The LE domain of **usherin** interacts with type IV collagen (panels A and B). Extracts of matrix from the indicated mouse tissues were (A) reacted with the fusion peptide comprising the LE-domain, immunoprecipitated with anti-GST antibodies, and the immunoprecipitate western blotted using anti-type IV collagen antibodies, or (B) directly immunoprecipitated with anti-type IV collagen antibodies and the immunoprecipitate western blotted using anti-**usherin** antibodies. The molecular weight markers are given in kilodaltons. The LN domain of **usherin** interacts with type I collagen (panel C). Extracts from the indicated tissues were reacted with the fusion peptide comprising the LN domain and immunoprecipitated with anti-GST antibodies. The immunoprecipitate was analyzed by western blot and probed with antibodies specific for type I collagen.

FIG. 9 is a Western blot illustrating the interaction of **usherin** with itself, possibly forming a suprastuctural network integrated into the basement membrane architecture. In panel A, the indicated fusion peptides were mixed with protein extracts from the eye, after removal of the lens (lanes 1, 3, 4, 6, 7, and 9) or the liver (lanes 2, 5, and 8) or with pre-immune serum (lanes 3, 6, and 9). The immunoprecipitate was analyzed by western blot probed with anti-**usherin** antibodies. Only the LN domain was capable of immunoprecipitating **usherin** from retinal extracts (lane 1). In panel B, purified fusion peptides were mixed in various combinations and crosslinked using dimethyl superimide (crosslinked mixtures are followed by an "X"). Products were resolved by PAGE, and stained with Coomassie blue. Arrows denote dimeric and trimeric crosslinked product.

L2 ANSWER 5 OF 11 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
DUPLICATE 4  
AN 2003:28147 BIOSIS  
DN PREV200300028147  
TI **Usherin expression** is highly conserved in mouse and human tissues.  
AU Pearsall, Nicole [Reprint Author]; Bhattacharya, Gautam; Wisecarver, Jim; Adams, Joe; Cosgrove, Dominic; Kimberling, William  
CS Boys Town National Research Hospital, 555 No. 30th St., Omaha, NE, USA  
kimber@boystown.org  
SO Hearing Research, (December 2002) Vol. 174, No. 1-2, pp. 55-63. print.  
ISSN: 0378-5955 (ISSN print).  
DT Article  
LA English  
ED Entered STN: 1 Jan 2003  
Last Updated on STN: 1 Jan 2003

L2 ANSWER 6 OF 11 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
DUPLICATE 5  
AN 2002:149106 BIOSIS  
DN PREV200200149106  
TI Localization and **expression of usherin**: A novel

basement membrane protein defective in people with Usher's syndrome type IIa.

AU Bhattacharya, Gautam; Miller, Caroline; Kimberling, William J.; Jablonski, Monica M.; Cosgrove, Dominic [Reprint author]

CS Boys Town National Research Hospital, 555 No. 30th St., Omaha, NE, USA  
cosgrove@boystown.org

SO Hearing Research, (January, 2002) Vol. 163, No. 1-2, pp. 1-11. print.  
CODEN: HERED3. ISSN: 0378-5955.

DT Article

LA English

ED Entered STN: 14 Feb 2002  
Last Updated on STN: 26 Feb 2002

L2 ANSWER 7 OF 11 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN  
DUPLICATE 6

AN 2001:351842 BIOSIS

DN PREV200100351842

TI **Expression, distribution, and integration of usherin:**  
A novel basement membrane protein defective in people with Usher syndrome type IIa.

AU Cosgrove, D. E. [Reprint author]; Bhattacharya, G. [Reprint author]; Kalluri, R.; Kimberling, W. J. [Reprint author]; Jablonski, M. M.

CS Genetics, Boys Town Nat'l Research Hosp, Omaha, NE, USA

SO IOVS, (March 15, 2001) Vol. 42, No. 4, pp. S654. print.  
Meeting Info.: Annual Meeting of the Association for Research in Vision and Ophthalmology. Fort Lauderdale, Florida, USA. April 29-May 04, 2001.  
Association for Research in Vision and Ophthalmology.

DT Conference; (Meeting)  
Conference; Abstract; (Meeting Abstract)

LA English

ED Entered STN: 25 Jul 2001  
Last Updated on STN: 19 Feb 2002

L2 ANSWER 8 OF 11 BIOSIS COPYRIGHT (c) 2005 The Thomson Corporation on STN

AN 2002:23328 BIOSIS

DN PREV200200023328

TI Distribution of **usherin** in humans and its effects on reproduction in people with usher syndrome type II.

AU Pearsall, N. A. [Reprint author]; Bhattacharya, G. [Reprint author]; Cosgrove, D. [Reprint author]; Wisecarver, J. L.; Kimberling, W. J. [Reprint author]

CS Genetics Department, Boys Town National Research Hospital, Omaha, NE, USA

SO American Journal of Human Genetics, (October, 2001) Vol. 69, No. 4 Supplement, pp. 651. print.  
Meeting Info.: 51st Annual Meeting of the American Society of Human Genetics. San Diego, California, USA. October 12-16, 2001.  
CODEN: AJHGAG. ISSN: 0002-9297.

DT Conference; (Meeting)  
Conference; Abstract; (Meeting Abstract)  
Conference; (Meeting Poster)

LA English

ED Entered STN: 26 Dec 2001  
Last Updated on STN: 25 Feb 2002

L2 ANSWER 9 OF 11 CAPLUS COPYRIGHT 2005 ACS on STN

AN 2000:790655 CAPLUS

DN 133:345575

TI Secretory **expression** systems for microorganisms using periplasmic chaperones and secretins of Gram-negative bacteria

IN Korpela, Timo; MacIntyre-Ayane, Sheila; Zavialov, Anton Vladimirovich; Battchikova, Natalia Vsevolodovna; Petrovskaya, Lada Evgenievna; Korobko, Vyacheslav Grigorievich; Zav'yalov, Vladimir Petrovich

PA Finland

SO PCT Int. Appl., 49 pp.

CODEN: PIXXD2

DT Patent

LA English

FAN.CNT 1

|      | PATENT NO.  | KIND | DATE     | APPLICATION NO. | DATE     |
|------|---|------|----------|-----------------|----------|
| PI   | WO 2000066756   | A1   | 20001109 | WO 2000-FI387   | 20000503 |
|      | W: AE, AL, AM, AT, AU, AZ, BA, BB, BG, BR, BY, CA, CH, CN, CR, CU, CZ, DE, DK, DM, EE, ES, FI, GB, GD, GE, GH, GM, HR, HU, ID, IL, IN, IS, JP, KE, KG, KP, LR, LS, LT, LU, LV, MA, MD, MG, MK, MN, MW, MX, NO, NZ, PL, PT, RO, RU, SD, SE, SG, SI, SK, SL, TJ, TM, TR, TT, TZ, UA, UG, US, UZ, VN, YU, ZA, ZW, AM, AZ, BY, KG, KZ, MD, RU, TJ, TM |      |          |                 |          |
|      | RW: GH, GM, KE, LS, MW, SD, SL, SZ, TZ, UG, ZW, AT, BE, CH, CY, DE, DK, ES, FI, FR, GB, GR, IE, IT, LU, MC, NL, PT, SE, BF, BJ, CF, CG, CI, CM, GA, GN, GW, ML, MR, NE, SN, TD, TG  |      |          |                 |          |
|      | FI 9901014  | A    | 20001105 | FI 1999-1014    | 19990504 |
|      | FI 109361   | B1   | 20020715 |                 |          |
|      | CA 2370436  | AA   | 20001109 | CA 2000-2370436 | 20000503 |
|      | EP 1173592  | A1   | 20020123 | EP 2000-922689  | 20000503 |
|      | R: AT, BE, CH, DE, DK, ES, FR, GB, GR, IT, LI, LU, NL, SE, MC, PT, IE, SI, LT, LV, FI, RO   |      |          |                 |          |
|      | NZ 515483   | A    | 20030926 | NZ 2000-515483  | 20000503 |
|      | AU 777246   | B2   | 20041007 | AU 2000-43003   | 20000503 |
|      | AU 2000043003   | A5   | 20001117 |                 |          |
|      | ZA 2001009231   | A    | 20021108 | ZA 2001-9231    | 20011108 |
| PRAI | FI 1999-1014  | A    | 19990504 |                 |          |
|      | WO 2000-FI387   | W    | 20000503 |                 |          |

RE.CNT 6 THERE ARE 6 CITED REFERENCES AVAILABLE FOR THIS RECORD  
ALL CITATIONS AVAILABLE IN THE RE FORMAT

L2 ANSWER 10 OF 11 FEDRIP COPYRIGHT 2005 NTIS on STN  
AN 2005:177436 FEDRIP  
NR CRISP 5R01DC004844-03  
TI **USHERIN: STRUCTURAL AND FUNCTIONAL ANALYSIS**  
SF Principal Investigator: COSGROVE, DOMINIC E; COSGROVE@BOYSTOWN.ORG, FATHER FLANAGAN'S BOYS' HOME, 555 NORTH 30TH STREET, OMAHA, NE 68131  
CSP FATHER FLANAGAN'S BOYS' HOME, BOYS TOWN, NEBRASKA  
CSS Supported By: NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION DISORDERS  
DB 2009 (/01/02)  
FYR 2004  
DE 2008 (/31/07)  
FU Noncompeting Continuation (Type 5)  
FS National Institutes of Health

L2 ANSWER 11 OF 11 GENBANK® COPYRIGHT 2005 on STN

LOCUS (LOC): CA757424 GenBank (R)  
GenBank ACC. NO. (GBN): CA757424  
GenBank VERSION (VER): CA757424.1 GI:25801463  
CAS REGISTRY NO. (RN): 550046-88-3  
SEQUENCE LENGTH (SQL): 255  
MOLECULE TYPE (CI): mRNA; linear  
DIVISION CODE (CI): **Expressed sequence tag**  
DATE (DATE): 27 Nov 2002  
DEFINITION (DEF): OD105G02\_T3.CRO OD Oryza sativa cDNA clone  
OD105G02\_T3.CRO similar to **usherin** [Rattus norvegicus], mRNA sequence.  
KEYWORDS (ST): EST  
SOURCE: Oryza sativa  
ORGANISM (ORGN): Oryza sativa

Eukaryota; Viridiplantae; Streptophyta; Embryophyta;  
Tracheophyta; Spermatophyta; Magnoliophyta; Liliopsida;  
Poales; Poaceae; Ehrhartoideae; Oryzeae; Oryza

NUCLEIC ACID COUNT (NA): 75 a 52 c 62 g 66 t

COMMENT:

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1201 W. Gregory Dr., Urbana, IL 61801, USA  
Tel: 2172655473  
Email: bohnertlab@life.uiuc.edu.

REFERENCE: 1 (bases 1 to 255)

AUTHOR (AU): Bohnert,H.J.; Borchert,C.; Brazille,S.; Brooks,J.;  
Eaton,M.; Ferrea,H. ; Kawasaki,S.; McCollough,A.;  
Michalowski,C.B.; Palacio,C.; Scara,G.; Wheeler,M.;  
Zepeda,G.R.

TITLE (TI): Functional Genomics of Plant Stress Tolerance

JOURNAL (SO): Unpublished (2000)

FEATURES (FEAT):

| Feature | Key    | Location | Qualifier  |
|---------|--------|----------|--|
| source  | 1..255 |          | /organism="Oryza sativa"<br>/strain="Pokkali"<br>/db-xref="taxon:4530"<br>/clone="OD105G02-T3.CRO"<br>/clone-lib="OD"<br>/tissue-type="roots"<br>/dev-stage="1 week"<br>/note="1 d 150mM NaCl" |

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61 gcagctgatc attataacaa tagcaagggtg caagccagct aacgaaatat aatcaagttt  
121 aacgaacctt tttgcactag tgaacacagc cggcttgcaa gttgtggatg gtttggtatg  
181 gaagcataga ggccgaccgc agtgagatga atggctgttt ttgacactgg gttaatgatt  
241 gtggacaaaa acctt

=>